

Acquired right atrial appendage to right ventricle accessory pathway in a lateral tunnel Fontan patient: Successful ablation via transconduit approach



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Introduction

The management of supraventricular arrhythmias after the Fontan procedure with catheter ablation represents a significant challenge. This can be due to the access problems that are created by the currently used modification of the procedure, which uses an extracardiac conduit, as well as to the complex anatomic substrate that can be created by a combination of congenital and acquired reasons. We report a case illustrating both of these aspects of the procedure and the potential solutions to the problems.

Case report

A 30-year-old man with history of complex congenital heart disease was referred for evaluation and treatment of preexcitation syndrome with frequent tachycardia that was resistant to medications. He was diagnosed with tricuspid atresia, hypoplastic right ventricle, ventricular septal defect, and pulmonary stenosis. He had undergone a pulmonary valvotomy at 3 days of age, placement of a modified Blalock–Taussig shunt at 6 months of age, and a Fontan operation of the atriopulmonary type at 4 years of age. Because of recurrent incisional atrial reentrant tachycardia (IART) that was resistant to multiple medications and catheter ablation, at 15 years of age, he underwent a partial resection of the anterior right atrium, an atrial septectomy, and the placement of an extracardiac Gore-Tex conduit from the inferior vena cava to the right pulmonary artery (RPA) with superior vena cava to RPA anastomosis. An

epicardial antitachycardia dual-pacemaker system was placed when he was 20 years old, and it was used several times to interrupt episodes of supraventricular tachycardia. At the same age, manifest preexcitation consistent with a right anterior accessory pathway was noticed for first time on his surface electrocardiogram (ECG) (Figure 1A). His tachycardias became typical paroxysmal supraventricular tachycardias (Figure 1B), while the episodes of IART were completely suppressed after the conversion to extracardiac Fontan. He was very symptomatic during the episodes of supraventricular tachycardia, requiring frequent hospital admissions for administration of adenosine and long-term treatment consisting of various medications including beta blockers, propafenone, and dronedarone. An attempt at catheter ablation through a retrograde transaortic approach using the Stereotaxis system (Stereotaxis Inc, St Louis, MO) was unsuccessful because of the inability to find any sites of early activation around the AV valve annulus. Mapping in the right atrial appendage was not attempted. The course of the extracardiac conduit was demonstrated by results of a computerized tomography scan of the chest (Figure 2A).

The patient was transferred to our hospital after a prolonged hospitalization that was prompted by recurrent supraventricular tachycardia and worsening renal and cardiac failure. After he received a detailed review of the risks and benefits of the procedure and granted informed consent, he was placed under general anesthesia. Access to the right atrium through the extracardiac conduit was guided by fluoroscopy and transesophageal echocardiography. After multiple attempts with different transeptal sheaths (Daig SR-0, Agillis, St Jude Medical, St Paul, MN) and a radio-frequency needle (NRG Transseptal Needle; Baylis Medical, Montreal, Canada) as well as a Brockenbrough transeptal needle, it was possible to advance initially a SafeSept wire (SafeSept, Pressure Products, San Pedro, CA) to the right and left atria, but the rest of the transeptal assembly (needle, dilator, and sheath) could not be advanced. A Terumo Glide catheter (Terumo Medical Corporation, Somerset, NJ) was used to exchange the SafeSept wire with an angioplasty wire.

KEYWORDS Accessory pathway; Extracardiac Fontan operation; Radiofrequency ablation; Transconduit puncture

ABBREVIATIONS AP = accessory pathway; ECG = electrocardiogram; IART = incisional atrial reentrant tachycardia; RPA = right pulmonary artery; VA = ventriculoatrial (Heart Rhythm Case Reports 2015;1:481–485)

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KEY TEACHING POINTS

- Acquired atrioventricular connections leading to Wolff-Parkinson-White syndrome may occur rarely after complex surgical repairs in Fontan patients.
- “Trans-septal” access across an extracardiac Gore-Tex conduit can be challenging, but it is feasible and may provide a superior approach for ablation compared with retrograde access.

After the newly created opening was dilated with a 5-mm coronary angioplasty balloon, it was possible to advance the

SR-0 sheath to the right atrium and have stable access for insertion of a mapping-ablation catheter. At baseline, the 12-lead ECG demonstrated ventricular preexcitation. The effective refractory period of the antegrade accessory pathway was short at 250 milliseconds. Atrial fibrillation was not induced. In order to avoid having a retrograde catheter in the arterial system and through the aortic valve for a long time, programmed atrial and ventricular stimulation was performed using the epicardial permanent pacing system. Narrow complex reentry tachycardia with 1:1 VA conduction was inducible initially with difficulty but subsequently very easily with ramped atrial pacing. The tachycardia was not tolerated hemodynamically, with systolic blood pressure being < 30 mm Hg, and therefore it was interrupted with

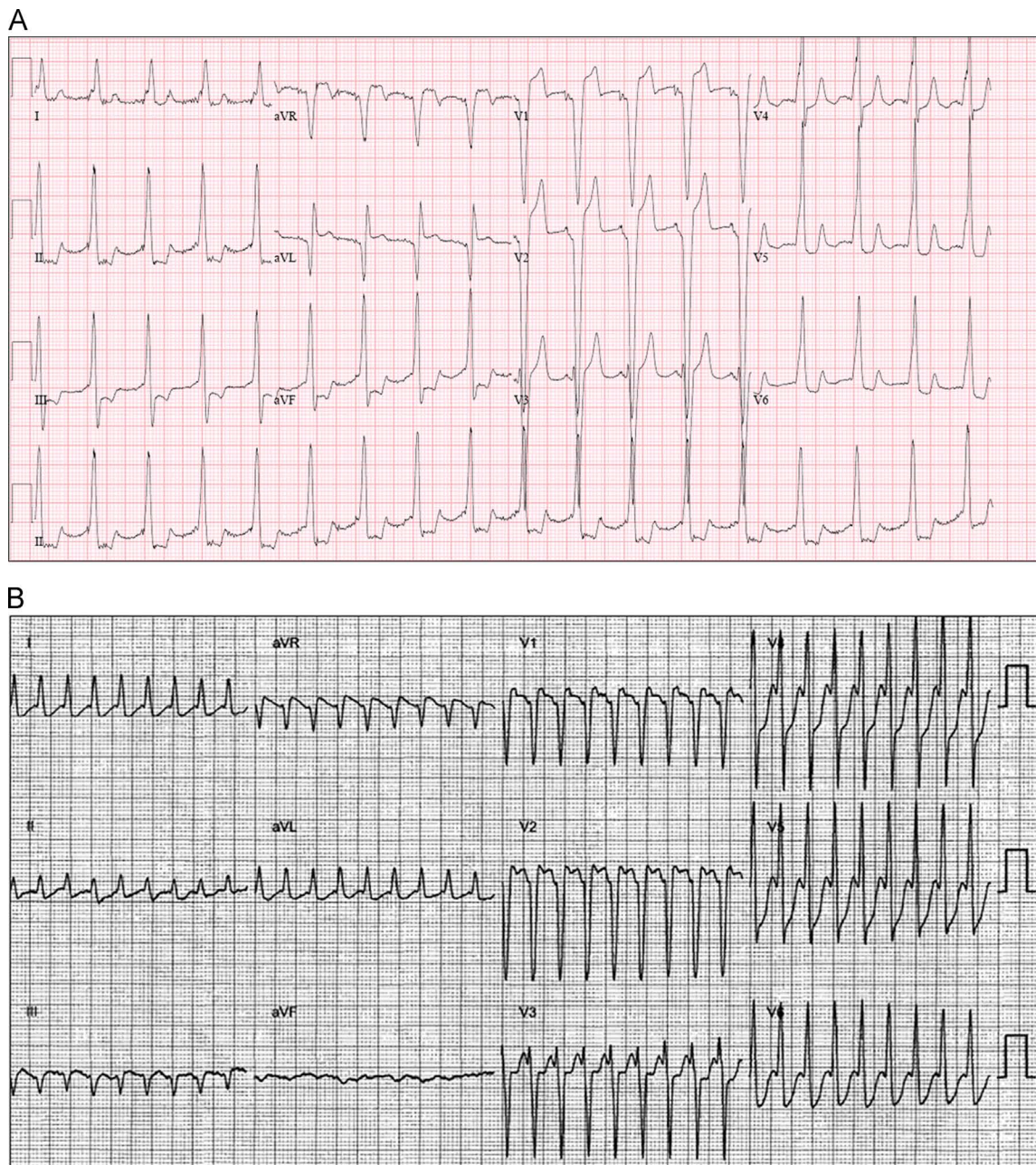


Figure 1 A: Baseline electrocardiogram with manifest preexcitation consistent with a right anterolateral accessory pathway. B: Supraventricular tachycardia (SVT) consistent with orthodromic reciprocating tachycardia.

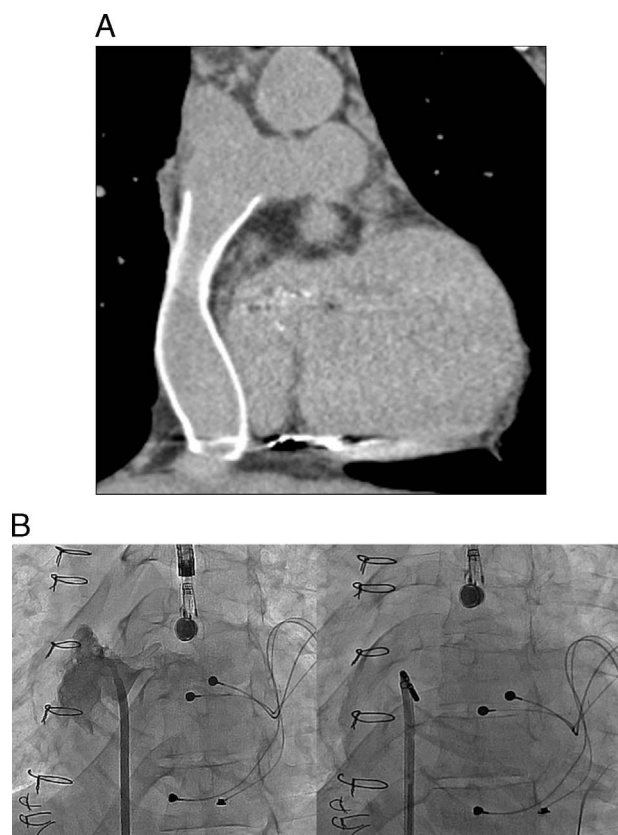


Figure 2 A: Results of computed tomography of the chest, demonstrating the extracardiac conduit adjacent to the right atrium. B: Right atrial appendage angiogram (left) and corresponding catheter position at the site of successful ablation of accessory pathway.

ventricular overdrive pacing through the epicardial pacemaker. After his hemodynamic condition was stabilized with dopamine and epinephrine infusions, it was possible to maintain tachycardia for several minutes with adequate blood pressures. It was not possible to perform diagnostic maneuvers such as placing a His-refractory premature ventricular beat, since all ventricular pacing was done through the epicardial permanent pacemaker. However, there was no VA block after adenosine administration during ventricular pacing, which was suggestive of using the accessory pathway for VA conduction. Mapping was initially performed during sinus rhythm, using the NavX system (EnSite NavX, St Jude Medical) and fluoroscopy guidance, searching for the earliest ventricular activation both around the right-sided atretic valve and around the mitral valve annulus. No early ventricular activation was seen in any annular location. While mapping more superiorly in the right atrial appendage during a spontaneous episode of tachycardia, early retrograde atrial activation was recorded. The catheter location within the right atrial appendage was confirmed using contrast injection through the long sheath (Figure 2B). The catheter was then readvanced, and activation mapping was performed during sinus rhythm with preexcitation and during orthodromic tachycardia with a 4-mm-tip radiofrequency catheter (Safire, St Jude Medical). All attempts to ablate the pathway were unsuccessful, despite

very early local electrograms. An 8-mm-tip catheter (Safire, St Jude Medical) was positioned at the base of the right atrial appendage (Figure 2B) and an early ventricular electrogram with a possible accessory pathway potential was found (Figure 3A). At this site, the accessory pathway was successfully ablated (Figure 3B) 20 seconds after onset of radiofrequency delivery. The maximum temperature was 60°C and the power that was delivered was 70 watts. Normal sinus rhythm without preexcitation was present after ablation. There was VA dissociation after adenosine administration during ventricular pacing. There was no recurrence of the pathway 1 hour after the successful lesion, and the catheter and the sheaths were removed. Transesophageal and transthoracic echocardiograms demonstrated an absence of pericardial effusion. The patient was discharged home 36 hours after the procedure. With no antiarrhythmic medications, he remains free of tachycardia, with no preexcitation on his ECG, 12 months after the ablation (Figure 3C).

Discussion

The presence of supraventricular tachycardia complicating the Fontan operation in patients with tricuspid atresia and other forms of functionally single ventricle has been well described. Most of these patients have IART.¹ The presence of typical accessory pathways in patients with tricuspid atresia has also been reported and surgical treatment of the accessory pathways simultaneous with Fontan operation has been described.^{2,3} Catheter ablation of the accessory pathway after the Fontan operation has also been reported.⁴ There have been a few patients described who developed surgically induced preexcitation after a surgically created anastomosis between the right atrium and right ventricle (Björk operation) for the treatment of tricuspid atresia.^{5,6} Our patient is unique, as his heart appears to have developed an unusual type of communication between the right atrial appendage and the right ventricle. Such a connection has been described before in patients with a structurally normal heart^{7,8} but not in patients with tricuspid atresia. We have no absolute proof that this pathway is acquired. However, several facts including the late development of manifest preexcitation following his last cardiac operation, as well as the lack of evidence of accessory pathway in his previous electrophysiologic study, suggest that this is a surgically acquired accessory connection. We hypothesize that after the extensive right atrial resection and resuturing, there was an inadvertent connection between the atrial appendage and the right ventricle. The site of the accessory connection was proven by right atrial angiography. This patient's symptoms changed significantly with the development of this accessory connection. His tachycardias became very severe, literally life-threatening, requiring multiple hospital admissions and drug administrations.

Regarding the technical aspects of the reported procedure, some points are of interest. The method for "transseptal" puncture through surgical baffles and conduits has been well

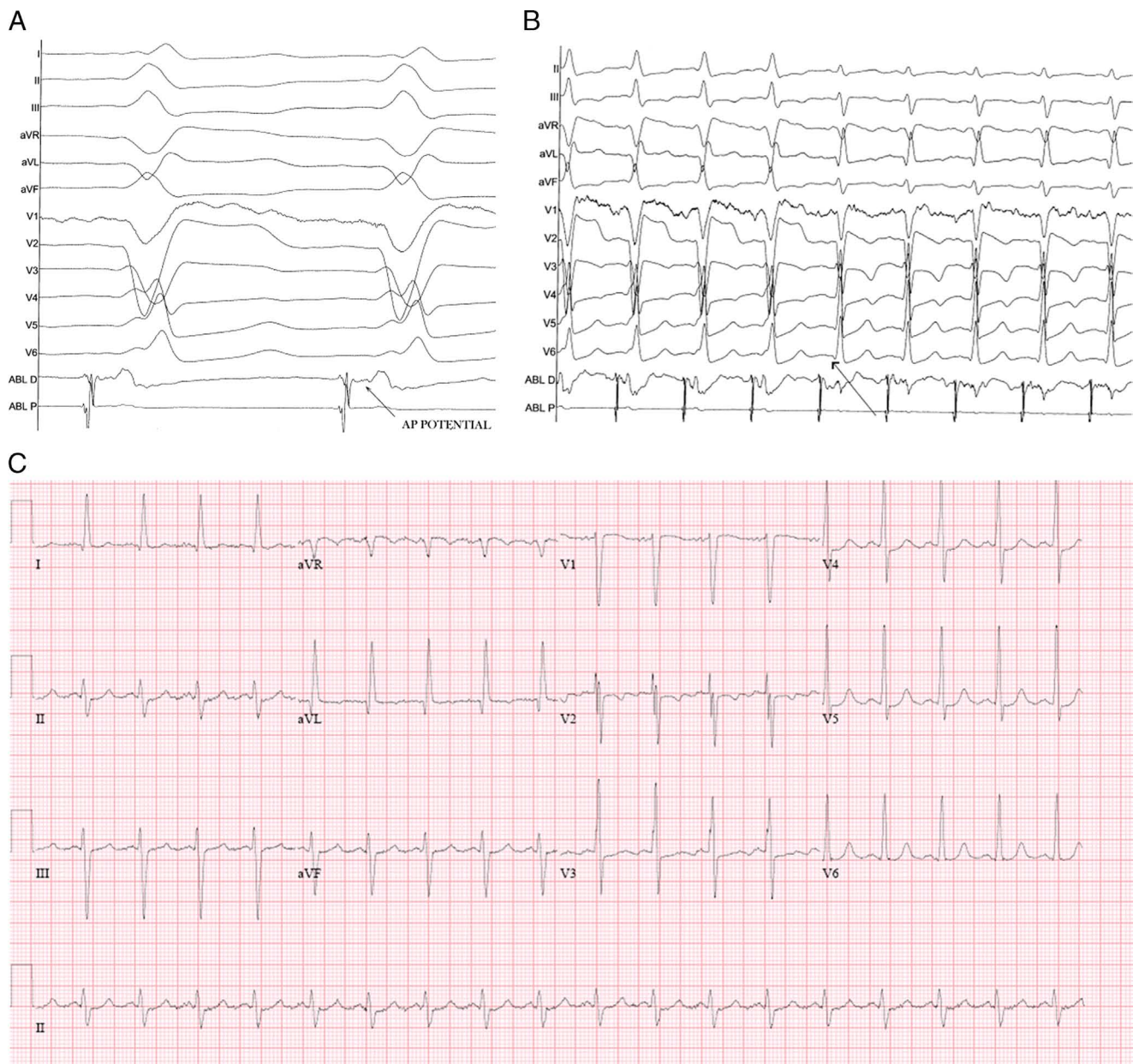


Figure 3 A: Successful ablation site with accessory pathway (AP) potential at the base of the right atrial appendage. B: Accessory pathway block during radiofrequency ablation. C: A 12-lead electrocardiogram, recorded 1 month post procedure, without preexcitation.

described.^{9,10} Puncture of the heavily calcified extracardiac conduit proved to be extremely difficult in our patient. Several transseptal needles failed, including a radiofrequency-powered needle and regular Brockenbrough transseptal needles. We describe a stepwise approach, using initially a SafeSept wire, to penetrate the wall of the conduit and the atrial free wall. Use of the SafeSept wire is an established method of crossing the atrial septum. The wire has a stiff end capable of perforating the septum, but it becomes curved as soon as it crosses, thus avoiding trauma to any nearby structures. After balloon dilation of the opening with a 5-mm coronary balloon, advancement of the 8.5F sheath and dilator was possible. Other authors have reported a new technique

consistent of snaring the dilator tip to improve stability and assist transbaffle puncture.¹¹

In terms of mapping and ablation, it is clear that exact localization of the accessory pathway by 12-lead ECG criteria is difficult in the absence of a true right AV valve annulus. The 12-lead ECG was suggestive of a right anterolateral accessory pathway. This suggested area is in fact where the pathway was located: not in the vicinity of the hypoplastic atretic AV annulus, which was at the “floor” of the right atrium, but between the base of the atrial appendage and the anterior RV free wall, which in a left lateral oblique view would correlate with a right anterolateral location. The pathway was probably epicardial, and therefore

ablation using common 4-mm-tip catheters was unsuccessful, because of the achievement of low power and low lesion depth. Ablation with an 8-mm-tip catheter resulted in delivery of high power leading to successful ablation of the accessory pathway.

Conclusion

An epicardial right atrial appendage to right ventricular connection was diagnosed in a patient with tricuspid atresia, after an extracardiac Fontan operation. We hypothesize that the accessory pathway was surgically created. The accessory pathway was successfully ablated using an approach from the extracardiac conduit to the right atrium and an 8-mm-tip ablation catheter.

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